

# CASE REPORTS

## ◀ The Surgical Management of Ruptured Omphalocele

## ◀ Renal Actinomycosis

### The Surgical Management of Ruptured Omphalocele

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**O**MPHALOCELE is congenital herniation of abdominal viscera through a deficiency in the rectus fascia. As it occurs at the base of the umbilical cord, the extrusion is covered by an exceedingly thin, translucent membrane composed of amnion and the underlying peritoneum.<sup>3</sup>

The contents of the sac, clearly visible through its wall, vary in proportion to the size of the fascial defect. Small intestine is almost always present, and not infrequently portions of the liver, stomach and colon. In extreme cases the hernia contains all the contents of the abdominal cavity. The integrity of the membrane is limited by lack of vascularity, and rupture is inevitable, although it may not occur for several days after birth.

The feasibility of surgical repair of omphalocele is now widely accepted, and there is conclusive evidence that the number of patients promptly referred for operation has risen sharply in recent years.<sup>1, 4, 9, 10</sup> As a result of increasing experience, cases formerly considered inoperable are being successfully treated, and at the same time the mortality rate has steadily declined.

Unfortunately, this encouraging trend does not apply to cases in which rupture of the sac has occurred. In a review of the literature, reports of only 15 cases in which operation was successful were found; six of the 15 cases were reported before 1900.<sup>1, 6, 8, 10, 11, 13, 16</sup> The average length of time between birth and operation was less than four hours.

Generalized fatal peritonitis develops rapidly in all untreated cases and is also the major cause of postoperative death in both ruptured and unruptured omphalocele. O'Leary and Clymer<sup>14</sup> in a review of 91 cases noted that the mortality rate rose from 21.4 per cent in cases in which operation was done within the first 12 hours, to 44.4 per cent for the group in which operation was carried out in the period between 12 and 24 hours, and to 61.6 per cent in cases in which operation was done more than 24 hours after delivery. Although the newborn is usually considered to be free of infection at the termination of a normal delivery, the degree of sterility is relative, not absolute—an important distinction that is seldom appreciated. It is, therefore, quite evident that the intact sac acts as a temporary barrier against infection. Moreover, it maintains the natural resistance of the peritoneum by protecting it from exposure and trauma. These two transient virtues of the membrane make possible the

relative success of present treatment in unruptured omphalocele.

However, when rupture is present at birth, or if it occurs in the delivery room, this advantage is missing. Exposure and peritoneal infection are concomitant and immediate. It is obvious that treatment of these conditions is as mandatory as repair of the congenital defect.

The purpose of this paper is to present an outline of procedure in ruptured omphalocele which fulfills these requirements and to report a case which is believed to be the first recorded of cure in a premature infant and the second in which an associated intra-abdominal anomaly was also corrected.

Since ruptured omphalocele is an emergency of the first magnitude, institution of treatment must be immediate and must be pursued with the least possible delay until completed. The importance of continuity of rigid sterile technique from birth to end of operation cannot be over-emphasized. Exposure of peritoneum in the delivery room instead of in the surgery is no excuse for contamination by unsterile material or personnel. Protection of the viscera from subnormal temperature and drying can be provided by use of moist warm saline compresses. The impulse to attempt reduction of the evisceration should be resisted, since taxis is seldom successful and causes severe trauma to the peritoneum.

A complication peculiar to small fascial defects is strangulation of the bowel produced by the expulsive action of lusty crying. If the intestine becomes cyanotic, the fascial orifice must be immediately extended—even though this permit further eventration. Finally, continuous administration of oxygen in a heated incubator is of distinct value while preparations are being made for operation.

These measures will minimize the hazards incident to rupture but they cannot be permitted to encourage procrastination. The only definitive treatment is surgical repair. There is no period of latency or incubation in peritonitis that may be relied on as a margin of safety, and unwarranted delay will defeat the most meticulous preoperative care.

General anesthesia is preferable to local infiltration. The risk involved is more than offset by less traumatic operation and decrease in operating time due to better exposure and relaxation. Before the bowel is replaced, there should be a careful inspection of the abdomen and intestinal tract. Concomitant anomalies are not infrequently encountered, and those incompatible with life, such as atresias and obstructions due to congenital bands, malrotation of the colon or imperforate anus, must obviously be corrected. Operation should not be prolonged, however, by unnecessary procedures such as correction of Meckel's diverticulum or appendectomy.

The repair of small defects is easily accomplished by approximating the peritoneum and posterior fascial sheath in the first layer, followed by anterior sheath, subcutaneous fat and the skin in the second layer. In dealing with larger

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defects it should always be remembered that closure with more than minimal tension may produce respiratory failure due to embarrassment of movement of the diaphragm, or circulatory failure secondary to pressure on the inferior vena cava. It is better to be content with closure of the subcutaneous fat and skin alone than to run the risk of these serious and usually fatal complications. And in the event that even this cannot be accomplished safely, dissection of the subcutaneous fat and skin must be carried as far laterally, superiorly and inferiorly as may be necessary to encompass the viscera without tension.<sup>5</sup> In such cases the risk of postoperative mechanical obstruction due to adhesions or necrosis of skin flaps without sufficient blood supply must be faced with the realization that they are complications inherent in a desperate situation.

If it has been impossible to close the fascial defect, there will remain a residual ventral hernia, often of immense size. Although grotesque in appearance and a source of concern to the parents, it does not interfere with the normal development of the infant. As time goes on, the abdominal cavity will increase in size and ultimately permit the viscera to occupy their normal confines. This may take many months, but secondary repair should be delayed until it has occurred and the surgeon is satisfied that he can successfully approximate the fascia.

#### CASE REPORT

A male infant weighing 4 pounds and 3 ounces was delivered two months before term with low forceps after a six-hour labor. The small intestine lay outside the abdominal cavity, having escaped through a defect approximately 1½ centimeters in diameter at the base of the superior aspect of the cord. The remnants of the ruptured sac could be identified at the skin edges.

Respiration was spontaneous and the baby was soon crying vigorously. It was then noted that the exposed segment of intestine was becoming quite cyanotic. An incision was immediately made through the full thickness of the abdominal wall, enlarging the defect by another 3 centimeters, which permitted the remainder of the small bowel, part of the stomach, the transverse colon and the cecum to eventrate. The viscera were covered with warm, moist normal saline compresses and the color of the segment which had been strangulated promptly returned to normal. The baby was then wrapped in warm sterile blankets and placed in a heated oxygen incubator while preparations were made for operation.

Approximately one hour after birth, operation was started under nitrous oxide ether inhalation anesthesia administered through an endotracheal tube with non-rebreathing valve. Constricted beneath a broad band near the base of the mesentery was a loop of small intestine in the shape of a three-leaf clover, with the central loop the longest of the three. Since neither patency nor continuity could be demonstrated beneath this band, it was tediously dissected free, which relieved the obstruction. The bowel was inadvertently opened during this procedure and immediately closed with an atraumatic chromic mattress suture, reinforced with a serosal stitch. Exploration revealed no further abnormalities. The abdomen was closed with two layers of interrupted fine cotton without causing undue tension.

The patient was returned to the oxygen incubator in good condition and given 50 cc. of 5 per cent glucose in Ringer's solution by clysis. Parenteral feeding, including blood transfusions, was continued along with oral feeding for the next two days, during which time the patient regurgitated frequently. Bowel movements occurred daily from the time of operation. By the fourth postoperative day most of the formula by mouth was being retained. A staphylococcal wound infection developed on the seventh day; it promptly

cleared when sutures were removed. The body weight reached a low of 3 pounds 13 ounces on the third day; then followed an uninterrupted gain to 4 pounds and 11 ounces on the 14th day.

At that time severe infectious diarrhea developed, and during the week required to bring it under control the body weight declined to within 1 ounce of the weight at birth. The patient was discharged on the 37th postoperative day, asymptomatic and eating well, with body weight 5 pounds 11 ounces. At eight months of age, the patient was healthy and energetic and without weakness of the abdominal wall. The body weight at that time was 20 pounds.

The incidence of omphalocele, intact or ruptured, occurring alone or with other anomalies, in stillborn or live births, operable or inoperable, is at present impossible to determine accurately for several reasons. Not the least of these is the confused nomenclature. The condition has been recorded as exomphalos, funicular hernia, amniocoele, congenital eventration and even as umbilical hernia. Therefore, it seems quite obvious that the vital statistics of public record must conceal many cases under even other diagnoses, especially if death ensued. The confusion of multiple names for this condition should be corrected by the universal adoption of the term omphalocele, as suggested by Ladd and Gross.<sup>6</sup>

It is indeed a rare anomaly and only some 400 cases have been reported to date in the world literature.<sup>2</sup> In large teaching clinics, where reasonable accuracy may be expected, the rate has been estimated to be from one in 4,000 to one in 11,000 births.<sup>2, 7, 12, 16</sup>

On the basis of these statistics, there would be approximately 40 cases a year in California alone—and it must be assumed that the vast majority of the patients die.

The number that might be saved will be determined only when obstetricians and surgeons are aware of the possibility of cure through proper preoperative care and prompt surgical intervention.

#### SUMMARY

Ruptured omphalocele is an urgent surgical emergency requiring continuous sterile technique and careful protection of the exposed bowel from birth until end of operation.

Concomitant anomalies incompatible with life must be recognized and corrected at time of operation.

The abdominal contents must be covered without excessive tension, even though a residual ventral hernia remains to be corrected at a secondary operation.

The successful treatment of a case complicated by prematurity and concomitant bowel obstruction due to congenital band is reported.

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(Cope in England and McWilliams in the United States, according to Davis).<sup>4</sup>

The oral cavity is the most likely portal of entry for the organism since the most common site of the disease is about the head and neck. Swallowed organisms may lodge in the cecum or appendix. Seven cases of renal actinomycosis following appendectomy have been reported. Gardiner<sup>7</sup> reported that 8 per cent of removed appendices contained colonies of actinomycetes.

Edwards<sup>8</sup> reported that Henrici was able only rarely to infect experimental animals with single injections of *A. bovis*. Repeated inoculations were required, suggesting that repeated exposure leading to sensitization was an etiologic factor.

Infection with other organisms may play a role in the initiation of the disease process, or in extension of it once it is present. Repeated sore throats or diseased tonsils have been noted in some patients prior to onset. Pneumonia or trauma to the chest have preceded pulmonary lesions in other instances.

## Renal Actinomycosis

### With Report of a Primary Case

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PRIMARY or solitary renal actinomycosis is a rare disease. Since Israel first reported a case in 1878<sup>1</sup> 31 others have been recorded. Only 15 of the reported cases appear to have been truly limited to the kidney. An additional case is reported here because of the rarity of the lesion and to emphasize that recent advances in therapy demand specific identification of the actinomycete if cure is to be obtained.

### ETIOLOGY

The cause of this disease is a saprophytic, facultative, anaerobic fungus. The colonies appear in the pus from lesions or are imbedded in granulation tissue and appear as irregular, yellow granules from 0.5 to 2 mm. in diameter. These are the so-called "sulfur granules." They may be soft and easily crushed or hard and calcareous. As observed microscopically, these granules have a central mass of debris, pus cells and degenerated fibers. Toward the margin is a dense network of interlacing, branched filaments. At the periphery are isolated, radiating filaments terminating in clubbed ends. The mycelial filaments are Gram-positive.

According to the classification of Weed and Bagenstoss<sup>2</sup> the human pathogen is called *Actinomyces israeli*. *A. bovis* is the anaerobic organism producing lumpy jaw in cattle. *Nocardia* is the name given the aerobic form of actinomycetes once thought to cause all cases in man and now incriminated in only 10 per cent of cases.<sup>2, 4</sup>

It was formerly thought that actinomycosis was contracted through ingestion or inhalation of contaminated straw, grains, or grasses, and this erroneous conception still is to be found in some present-day textbooks. It has been proved that *A. israeli* lives as a harmless saprophyte in the mouths, tonsils, and intestinal tracts of normal human subjects. Davis<sup>4</sup> stated that Naeslund cultured pure colonies of the organism from the mouths of healthy persons. These colonies, on injection, produced the disease in guinea pigs. Two cases have been reported in which actinomycosis followed human bites by persons without evidence of the disease

### PATHOLOGY

The term primary renal actinomycosis is used here in the same sense that renal tuberculosis is termed primary. That is, it must be assumed that an earlier lesion existed somewhere in the body from which the kidney became infected. The patient probably was symptomless at the time of this early lesion and demonstrable residual traces are absent. That it must have existed, however, is indicated by the presence of the disease in the kidney.

The characteristic lesions are chronic abscesses as a result of progressive penetration and destruction of tissue. Tissue reacts to the invading parasite by the formation of nodules of granulation tissue rich in vessels and cells. The centers of these nodules then break down by a process of lipoid degeneration and become filled with leukocytes, debris, and sulfur granules. In the wall of granulation tissue about the abscesses are many mononuclear and occasional giant cells. As the lesion ages, pronounced formation of connective tissue replaces most of the granulation tissue. Thus the lesion of actinomycosis is a chronic, suppurating granuloma. The lesions vary in size from that of a pinhead to that of a grapefruit.

Extension from the primary site is usually by direct spread to tissue and along fascial planes. The lymphatic system is almost immune and the lymph nodes do not react and enlarge. The skin is involved late in the process, as it offers great resistance. Muscles and nerves may be invaded or pushed aside. Bones may be superficially involved by direct continuity. Rarely, hematogenous spread occurs and then any organ may be involved. Primary renal actinomycosis is initiated in this manner. Secondary renal lesions occur when the kidney is involved contiguously.

Grossly, a variety of pathologic changes may be observed in the infected kidney. When the lesion is minimal, only one small area is affected. On cut surface of the kidney a pyramidal area of granulation and scar tissue may be observed, the apex pointing toward the renal pelvis in the region of a renal papilla indicating the hematogenous origin of the infection. Within this may be seen yellowish streaks or granules, and abscess cavities of varying size which may also contain the yellow sulfur granules. The granuloma may extend through the capsule to invade the perinephric fat. Frequently a perirenal abscess is produced. When involvement is more diffuse, the entire kidney may be converted into a suppurating, granulomatous mass. Subsequently, multiple sinus tracts discharge to the skin. If the involvement begins in the lower pole and spreads to include and obstruct the

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